Idebenone increases mitochondrial complex I activity in fibroblasts from LHON patients while producing contradictory effects on respiration

Titre	Idebenone increases mitochondrial complex I activity in fibroblasts from LHON patients while producing contradictory effects on respiration
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Résumé en anglais	BACKGROUND: Leber's hereditary optic neuropathy (LHON) is caused by mutations in the complex I subunits of the respiratory chain. Although patients have been treated with idebenone since 1992, the efficacy of the drug is still a matter of debate. METHODS: We evaluated the effect of idebenone in fibroblasts from LHON patients using enzymatic and polarographic measurements. RESULTS: Complex I activity was 42% greater in treated fibroblasts compared to controls (p = 0.002). Despite this complex I activity improvement, the effects on mitochondrial respiration were contradictory, leading to impairment in some cases and stimulation in others. CONCLUSION: These results indicate that idebenone is able to compensate the complex I deficiency in LHON patient cells with variable effects on respiration, indicating that the patients might not be equally likely to benefit from the treatment.
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